CASE REPORT

Subtrochanteric fractures in osteopetrosis; report of two cases with conservative and operative management

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Abstract
In osteopetrosis subtrochanteric area is especially vulnerable to get fractured as it is an area of concentrated stress. We describe two cases of autosomal dominant osteopetrosis with subtrochanteric fractures. In one case upper tibial pin track infection prompted us towards a conservative management with a successful outcome in a time span of 25 weeks. The second case was directly treated by dynamic hip screw fixation though with great difficulty but was complicated by poor callus formation and loosening of implant. Re intervention was done with change of implant and bone grafting with subsequent fracture union. Both the cases experienced a second subtrochanteric fracture in their opposite hips after an average time period of 6 years and were managed operatively. Our experience with these two cases makes us to believe that there can be no clear cut protocol for the management of subtrochanteric fractures in osteopetrosis. Conservative management can still be considered when operative treatment is not an option. On the other hand, operative treatment is associated with considerable difficulty and complications. We thus have to weigh our options accordingly.

Key words: Osteopetrosis, subtrochanteric fracture, conservative management, operative management

INTRODUCTION
Osteopetrosis (marble bone disease) is a descriptive term that refers to a group of rare, heritable disorders of the skeleton characterized by increased bone density on radiographs. German Radiologist Albers-Schoenberg was the first person to describe the condition in 1904 (1). This disease was referred to as “osteopetrosis” by Karshner in 1926 (2). Even though bone has a dense sclerotic appearance on radiographs, it is quite weak, fragile and prone to fractures (3,4,5). Subtrochanteric area is quite susceptible to fractures in osteopetrosis because this is an area of stress. Osteopetrotic fractures are a treatment challenge and most of the literature concentrates on the difficulties of operative management and associated complications (3-15). We present two cases of autosomal dominant osteopetrosis with subtrochanteric fractures in an adult age group with an average of 6 year follow up. With one case managed conservatively and the other one by an operative intervention, the response to the two modalities of treatment in the two cases is described.

CASE 1
A 20 year old patient reported to our emergency department in March 2002. She gave a history of trauma right hip after sustaining a fall on a hard floor. She was a known case of autosomal dominant osteopetrosis which had been diagnosed earlier. Plain X ray revealed a right subtrochanteric fracture. The patient was admitted to the hospital for tibial pin traction to await surgery. The patient’s condition was complicated by a pin track infection which developed a week after her admission despite being given an adequate antibiotic prophylaxis. Her condition alerted us against an invasive procedure and a conservative management was decided upon. She was put on a below knee skin traction for five weeks followed by traction in Thomas splint for a period of 20 weeks. Even though the fracture united in a slight varus...
position, she returned to her normal routine activities. Her pin track infection got complicated into chronic osteomyelitis which was subsequently treated and resolved over a period of one year (figure 1). Patient again reported to us in February 2009 with a fresh trauma to left hip. X ray pelvis AP view showed fracture subtrochanter left hip with a united fracture on right side (figure 2). This time fracture was fixed by dynamic hip screw fixation (figure 3).

CASE 2

In May 2004, a 34 year old male, known case of osteopetrosis was received in our emergency as trauma left hip after a fall. X ray pelvis revealed a fracture subtrochanter left hip. The patient was admitted for management. Upper tibial pin for traction was placed with difficulty. Patient was put on skeletal traction for a period of seven days following which Dynamic Hip Screw fixation was done. Even after using power drills, difficulty during drilling was encountered resulting in breaking of three drill bits. Patient was discharged and put on a follow up. X ray pelvis at 8 month revealed poor callus formation with loosening of implant. A second intervention was decided. The implant was exchanged with a large ten hole DHS plate. Fracture site was grafted with the bone harvested from left iliac crest. The fracture united and patient was back to his normal routine (figure 4). Patient again reported to us in April 2009 with fresh trauma to right hip. X ray pelvis revealed a right sided subtrochanteric fracture (figure 5). After a period of five days on pin traction, DHS fixation was done but with a lesser difficulty.

DISCUSSION
Osteopetrosis is caused by failure of osteoclast differentiation or function. This results in failure of bone remodeling (16). Callus formed after a fracture in osteopetrotic bone has less number of osteoclast and lacks a well organized Haversian system (6). The severity of osteopetrosis ranges from fatal in infancy to asymptomatic in adults with a normal life span. The more fatal forms are inherited as autosomal recessive while the mildest forms observed in adults are inherited in an autosomal dominant manner. Depending on the location of osteosclerosis, autosomal dominant has been divided into two subtypes (17, 18). While first type involves the cranial vault, the second type involves spine and pelvis. It is the type II which is known as Albers-Schoenber disease. There have been at least 10 genetic mutations described which are linked to causation of osteopetrosis. In Autosomal dominant variety, fractures involving subtrochanteric areas are common. Both operative and nonoperative modalities of treatment have been used to treat such fractures. Most of the literature written about osteopetrosis stress on the difficulties encountered while operating and associated complications. Armstrong et al treated peritrochanteric fractures using screw plate or a nail plate (3). He described placement of fixation devices as difficult and in one patient the reamer for the screw plate was destroyed. Ashby while treating a peritrochanteric fracture with a zickel nail described it as difficult, surgery taking 6 hours to complete, with an extremely hard bone to drill (7). Milgram and Jasty found it difficult to place a holt nail plate while treating a peritrochanteric fracture (5). Yang et al (8) failed in placing a jewett nail in a 21 year old female with a peritrochanteric fracture and Kleinberg (9) broke plate while fixing the same fracture. P. Birmingham (19) studied the case reports and small case series on operative treatment of peritrochanteric fractures and neck fractures in osteopetrosis (four femoral neck fractures and 21 peritrochanteric fractures). He found a 12% nonunion rate and a 12% infection rate. In the peritrochanteric group, the rate of hardware failure was 29%, rate of reoperation 29%, and there was a 14% incidence of periprosthetic fracture.

Nonoperative treatment for osteopetrotic femoral fractures has been described by Alexander (14), Hasenhuettl (15), Armstrong et al (3), Birmingham (19). They have used a wide variety of techniques like plaster cast, Russel traction, Hip spica and even a simple traction . The complications they report include pseudoarthrosis, nonunion, and coxa vara. Birmingham describes Coxa Vara as the most common complication (31%) of Nonoperative treatment (19). As per the case reports he studied, 30 patients of 35 achieved union with no reported delays and complications.

Our experience with the two cases was varied. Case 1 though managed conservatively was non ambulatory for a long time which in our view is a major disadvantage of this method. The fracture united in a slight varus position but the functional outcome was fine. Operative intervention was done in case 2. This method of intervention was complicated by poor callus formation, loosening of implant. Union was achieved by re intervention, by implant exchange and bone grafting. In both the patients upper tibial pin was placed with great difficulty and while operating case 2 three drill bits broke in the bone. We again got a chance of operating these two cases after an average time of 6 years. We experienced lesser difficulty operating them again. We believe that the osteopetrotic bone behaves differently at different ages. Probably an osteopetrotic bone in a younger age is harder and more likely to undergo complications such as osteomyelitis but we don’t have a conclusive proof to support this hypothesis.

Conservative management can still be considered when operative treatment is not an option. On the other hand, operative treatment is associated with considerable difficulty and complications. We thus have to weigh our options accordingly.

Conflict of interest statement;
We certify that there is no actual or potential conflict of interest in relation to this article.

REFERENCES